

8. DEFECTS OF THE DIAPHRAGM (F)

Diaphragmatic hernia is a not uncommon cause of death in the first few days of life, but is found much less frequently in the stillborn. The most common type is due to an opening on the dorso-lateral aspect of the diaphragm attributed to failure of completion of separation of the pleural and abdominal cavities by the plural peritoneal folds. These openings are posterior to that portion of the diaphragm formed by the septum transversum, which had developed in the cervical region and descended caudally as the embryo developed. A majority of these hernias are left-sided and those occurring on the right side are usually smaller. Severe cases are often described as absence of one leaf of the diaphragm.

Other forms of hernia are much less common, perhaps the most frequent being a weakening of the diaphragm, again most commonly in the left side, resulting in stretching and pressure upwards by the abdominal contents on the heart and lungs. Almost invariably there is aplasia of the lung on the side of all such hernias and this cannot be regarded as an independent defect. Malformation of the heart is also common and some of these malformations appear to be due to a contiguity effect while others

are probably related to the disturbance of development of the primitive mesentery.

CASES WHERE DIAPHRAGMATIC HERNIA WAS THE ONLY DEFECT RECORDED

In all there were 47 single-born infants where a defect of the diaphragm was the only one recorded in the study. In view of the small numbers no general table appears to be justified but the details will be found in the Basic Tabulations by Centres booklet. Of these 47 cases, 25 were in males, 22 in females. Thirty-nine were stillborn or died in hospital. Deaths from this cause thus represent about 0.5% of all hospital deaths. The site of the hernia was recorded in only 20 of these cases and of these 17 were on the left side. There were no cases of diaphragmatic hernia in twins.

DIAPHRAGMATIC HERNIA AS ONE OF MULTIPLE DEFECTS

In the N or multiple group there were 10 cases (4 males and 6 females). The types of associated malformations in these cases are shown in Table 8.1.

TABLE 8.1
DIAPHRAGM DEFECTS IN SINGLE BIRTHS, OCCURRING WITH OTHER DEFECTS IN THE N GROUP

Centre	Sex	Survival	Type of defect of diaphragm	Other defects
Melbourne 1	F	SB	Defect of left lobe	Tracheo-oesophageal fistula
Melbourne 2	M	SB	Defect of left lobe	IASD
Czechoslovakia	M	LBA	Diaphragmatic hernia (L)	IVSD
Hong Kong	F	LBD	Diaphragm defect (L)	Malformation of upper limb (L) (NFS)
Hong Kong	F	LBD	Diaphragm defect (R)	IVSD; mitral atresia
Mexico 2	F	LBD	Absence of leaf of diaphragm (NFS)	Cranium bifidum
Belfast	M	LBD	Hiatus hernia	HL/CP (NFS)
Manila	M	LBD	Diaphragmatic hernia (NFS)	HL/CP (L); rudimentary genitalia
Madrid	F	LBD	" Abnormal " herniated diaphragm	Rudimentary sternum; CHD (NFS); rudimentary fingers (R); thymus adherent to pericardium
Ljubljana	F	LBD	Diaphragmatic hernia (NFS)	IVSD; ? Bonnevie-Ullrich syndrome